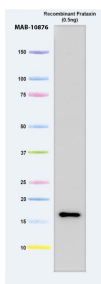


<b>Cat. No:</b>	MAB-10876
<b>Conjugate:</b>	Unconjugated
<b>Size:</b>	100 ug
<b>Clone:</b>	STR23
<b>Concentration:</b>	1mg/ml
<b>Host:</b>	Ms
<b>Isotype:</b>	IgG1,k
<b>Immunogen:</b>	Human frataxin (full length mature form, residues 56-210)
<b>Reactivity:</b>	Hu
<b>Applications:</b>	Western blotting , Immunocytochemistry, Flow Cytometry , IHC(P)
<b>Purification:</b>	Purified

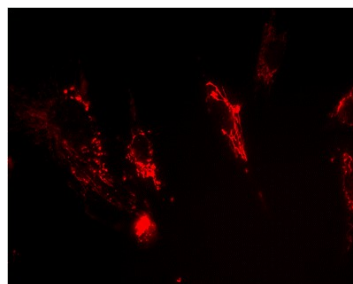
**Background:**

Frataxin is a mitochondrial protein that is approximately 17 kDa in size in its monomeric form. The gene encoding frataxin in humans lies on chromosome 9. The codon GAA is repeated 100-1700 times in both copies of this gene in the majority of patients having Friedreich's Ataxia (FRDA). FRDA is an autosomal recessive disorder causing neurodegeneration and cardiomyopathy. FRDA patients show low expression of frataxin. Frataxin has been implicated in mitochondrial iron homeostasis although the mechanism by which this protein is involved in iron metabolism is still not clear. Deletion of the frataxin gene in yeast results in iron accumulation in the mitochondria and loss of respiration. Increased mitochondrial iron levels have also been observed in FRDA patients. Recombinant human frataxin, both in its monomeric form and in a polymeric form, has been shown to bind iron in vitro.

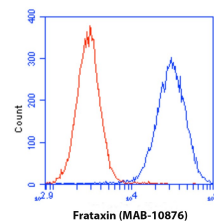
**Form:** Liquid



Recombinant Human Frataxin (lane 1) detected with (MAB-10876) anti-Fratxin antibody



Immunocytochemistry analysis using MAB-10876 at 1 ug/mL staining Frataxin in normal Human Fibroblasts (MRC5) (fixed, permeabilized) followed by an AlexaFluor® 594-conjugated-goat-anti-mouse IgG1 isotype specific secondary antibody (2 µg/ml).



HL-60 cells were stained with 2 µg/mL Frataxin antibody (MAB-10876) (blue) or an equal amount of an isotype control antibody (red) and analyzed by flow cytometry.

**Suggested Working**

6-10 µg/ml for Western blotting (a chemiluminescent detection system is highly recommended),

1 µg/ml for Immunocytochemistry , 2 µg/ml for Flow Cytometry, 20 ug/mL Immunohistochemistry

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